

Clinical Variations of Cardiac Sarcoma

To the Editor:

Malignant cardiac tumours constitute a group of tumours with a very low incidence but high mortality that are difficult to diagnose, primarily because of their various forms of presentation and symptoms. We present 3 cases of cardiac sarcomas that clearly represent the variety of their clinical presentation and the respective chain of events of this type of tumours.

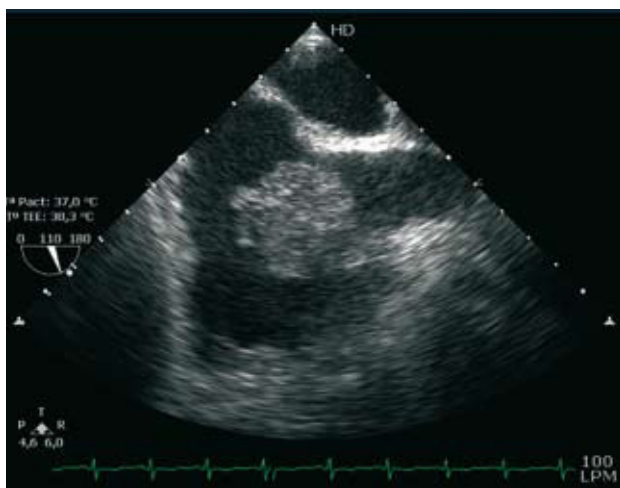


Figure 1. Image of the transoesophageal echocardiogram. View of the chambers where a mass with irregular edges can be observed with a small thrombus adhered to the wall of the right atrium.

Case 1. Twenty-seven-year-old male attended to in the emergency room for pleuritic chest pain, low grade fever, and asthenia. The initial physical exploration was normal. He was admitted with suspected diagnosis of acute viral pericarditis. After carrying out transthoracic echocardiography (TTE) and transoesophageal echocardiography (TEE), a large irregular-shaped mass was found in the interatrial septum that infiltrated the right atrium, the tricuspid ring and the base of the aorta; it produced a subtotal stenosis of the superior vena cava and severe pericardiac effusion. A biopsy of the mass was taken by thoracotomy. The pathological anatomy confirmed the diagnosis of a cardiac angiosarcoma. Chemotherapy was initiated with intravenous taxol. The patient passed away 6 months after the diagnosis.

Case 2. Forty-year-old male with symptoms of partial simple epileptic crises. An electroencephalogram (normal) and a computerized cerebral tomography were carried out that showed a frontal mass with a large perilesional edema, indicating a cerebral metastasis of an unknown primary tumour. The thoraco-abdominal tomography showed multiple intrapulmonary masses of a small size compatible with metastases and a mass in the right atrium. The TEE showed a large mass with irregular edges in the right atrium without invading any other structures (Figure 1). With a biopsy of one of the pulmonary masses, an anatomopathological diagnosis was made of metastases of a cardiac angiosarcoma (Figure 2). Chemotherapy was initiated with intravenous taxol, but the patient passed away 2 months after the diagnosis was made.

Case 3. Fifty-nine-year-old women with symptoms of asthenia, low grade fever, and dyspnea from

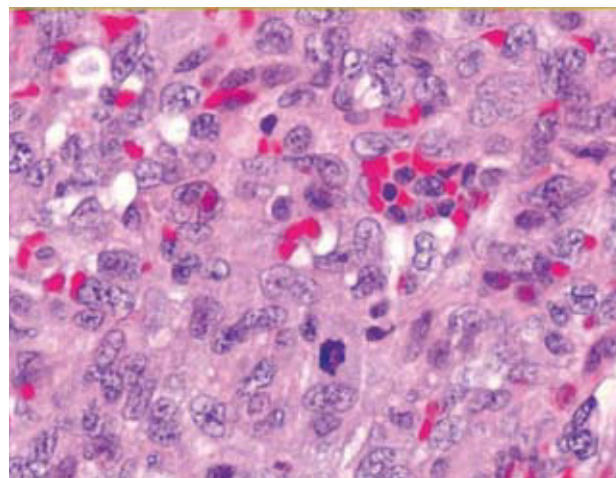


Figure 2. Microscopic vision of a pulmonary metastatic nodule of the angiosarcoma. Hematoxylin-eosin stain where vascular structures and neoplastic cells are observed in different phases of mitosis.

moderate exercise of 1 week of evolution. The physical exploration showed a systolic murmur III/IV from a pulmonary nucleus. She was admitted to investigate a probable valvulopathy. After little more than 24 hours, she suddenly died from a cardiorespiratory failure. A large tumour in the principal trunk of the pulmonary artery that totally occluded the pulmonary valve was found at autopsy. The microscopic study showed a highly malignant sarcoma.

Malignant cardiac tumours are a rare entity. Of these, the most prevalent primary malignant tumours are the sarcomas, fundamentally the angiosarcoma,¹ as taken from the National Registry of Cardiac Tumours.²

The symptoms are variable and generally infrequent, and therefore the diagnosis is usually made by coincidence, as specified in one of the last reviews on this topic by Burke et al.³ In our cases, we found local invasion of the pericardium, distant metastases and invasion of large blood vessels. Each one of the cases presented totally different symptoms, but a similar etiological substrate.

The diagnosis should be started with TTE and TEE⁴ that enable the determination of the localisation of the mass, and the repercussion in the cardiac hemodynamics. Computerised tomography and magnetic resonance imaging make it possible to know the degree of infiltration of the myocardium and of adjacent structures.⁵

Due to its increased aggressiveness, this type of tumours has a poor prognosis. Survival varies between 6 and 12 months,⁶ although there have been exceptional cases of 3 years of survival.⁷

Total surgical resection of the tumour is hard to apply, as 80% present invasion of neighbouring structures at the time of diagnosis. Heart transplants have been considered in patients without distant metastases. However, improvement of survival times have not been demonstrated in the cases that have been carried out.⁶

Given the aggressiveness, the lack of obvious symptoms and the high rate of mortality of this type of tumour, further understanding is needed. The series that we present clearly shows the extreme variability of the localisations that they can present and, consequently, of the symptoms that they cause.

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